Operative Techniques in Thoracic and Cardiovascular Surgery

Surgical Therapy for Anomalous Aortic Origin of the Coronary Arteries

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Anomalous aortic origin of a coronary artery (AAOCA) from the incorrect sinus of Valsalva is a rare congenital cardiac defect that is associated with increased risk of sudden death and cardiac morbidity.[1-3] The incidence of this defect approximates 0.17% in autopsy series, and 0.1 to 0.3% in patients undergoing catheterization or echocardiography.[1-4] In adult patients referred for coronary angiography, the prevalence of all congenital coronary artery anomalies is 0.6 to 1.3%.[5,6] The most common coronary anomaly, with an incidence of 0.37 to 0.6%, is that in which the circumflex coronary artery arises from the right sinus or the right coronary artery.[7,8] The anomalous circumflex has little clinical significance when found in isolation. The next most common and pathologically significant anomalies are the right coronary artery from the left sinus of Valsalva (ARCA), and the left main coronary artery from the right sinus of Valsalva (ALMCA). The natural history of these anomalous coronary arteries is not well described and currently must be inferred from autopsy series and anecdotes. It appears that risk is most pronounced in young, competitive athletes.[1,2] Coronary anomalies are the second leading cause of cardiac death in young athletes. In post-mortem study, 57% of the 49 patients with ALMCA, and 25% of those with ARCA, died suddenly.[9] Increased risk of sudden death associated with exercise is well described with both ALMCA and ARCA.[2,3,10]

Anomalous aortic origin of the coronary artery may be associated with cardiovascular symptoms most often referable to ischemia. These include angina, arrhythmia, syncope, and sudden death.[11] Because the prevalence of the defect is not known, it is currently impossible to define the prevalence of symptoms. The patient is not uncommonly asymptomatic. The diagnosis is made serendipitously during provocative testing for other cardiovascular disease with catheterization or echocardiography.[4,6]

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In children, the diagnosis is most frequently made with echocardiography, which is usually sufficient to characterize the defect and to guide surgical repair.[12] Computed tomography angiography and cardiac magnetic resonance imaging are useful, especially in older patients in whom echocardiographic windows are inadequate, and to differentiate between intra- and extramural course of the coronary. Coronary angiography is a very useful technique to detect an interarterial course but is not very useful to distinguish between intra- and extramural course.[13] We tend to reserve cardiac catheterization for patients in whom the diagnosis is in question, or for adult patients with risk factors for atherosclerotic coronary artery disease.

Indications for Surgery

In patients with AAOCA and symptoms referable to coronary ischemia, there is no debate regarding indication for operative repair. However, the management of the asymptomatic patient with AAOCA remains somewhat controversial. The risk of sudden death or cardiac ischemia must be weighed against the risk of the operation. In asymptomatic patients with ALMCA, there seems to be consensus that surgical intervention is indicated to prevent the risk of sudden death. Because the risk of sudden death in patients with ARCA is significantly less than with ALMCA, the decision for elective surgical intervention is more difficult, but, in our opinion, still warranted. In asymptomatic patients with ARCA, our recommendation is to delay elective surgical repair until late puberty or approximately 10 years of age. This recommendation is based on data that suggest that cardiac-related symptoms and sudden death in children with ARCA are rare before adolescence.[14] If surgical repair is declined or deferred, avoidance of strenuous physical activity and competitive athletics is often prescribed. However, this is not necessarily protective, because in more than half of patients with ARCA and sudden cardiac death, death was not associated with exercise.[9]

It must be said that there is considerable debate regarding indications for intervention; ongoing efforts should help establish a more unified approach in this group of defects.

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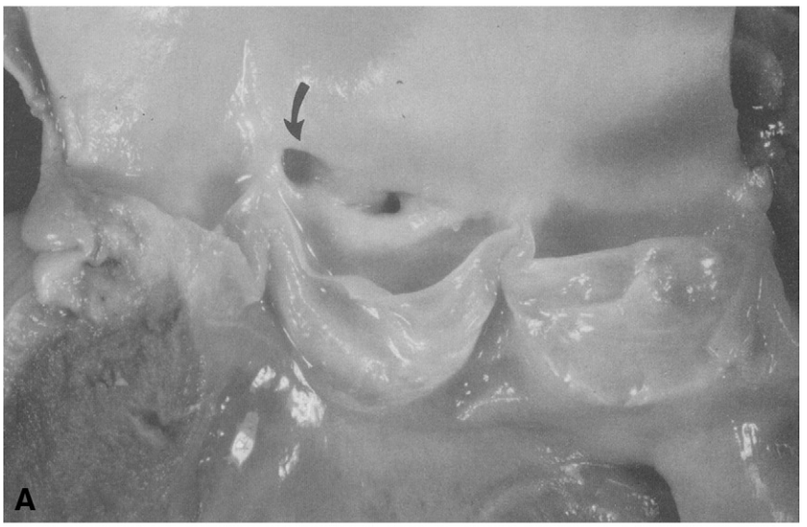
Operative Technique



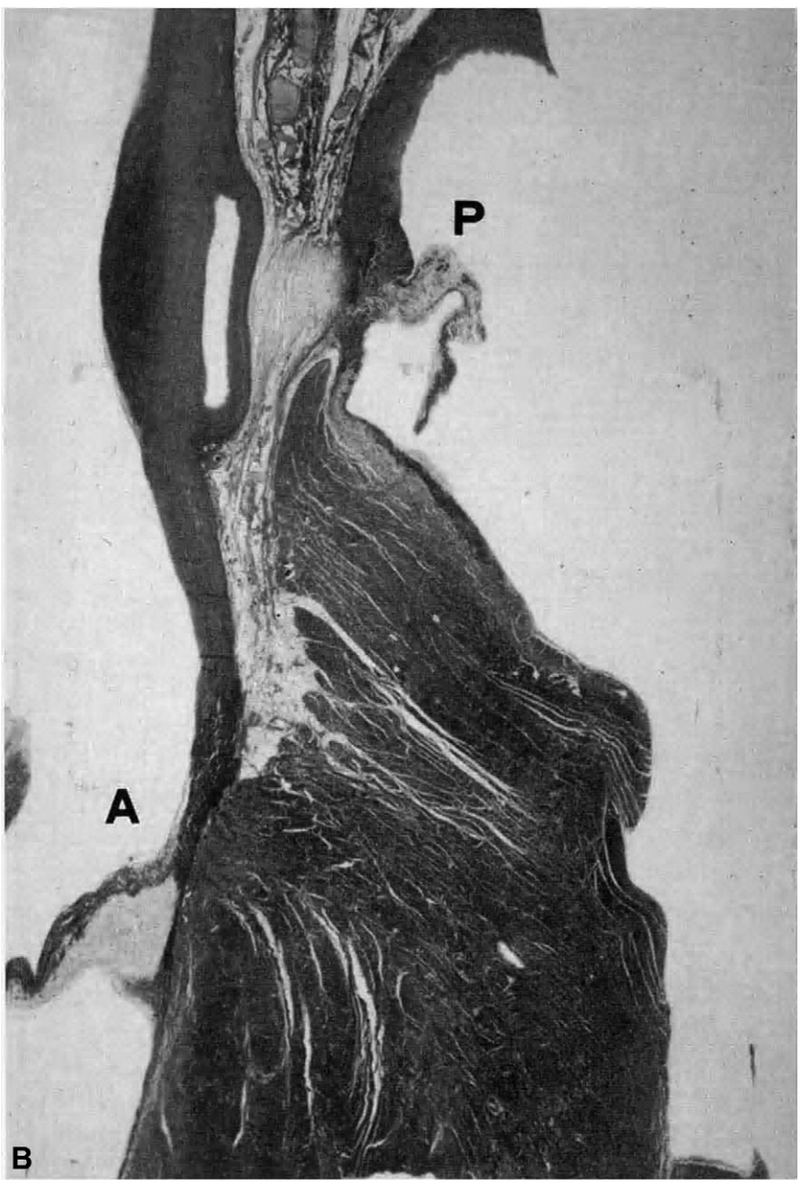


Figure 1 (A) Autopsy specimen of a 32-year-old woman who died suddenly while jogging. The arrow shows an ALMCA from the right sinus of Valsalva, originating at the level of and immediately to the right of the intercoronary commissure. There is demonstrated a flap or slit-like orifice of the anomalous coronary. Pathologic studies have identified several features that are believed to underlie symptoms of ischemia.[21] The importance of the flap or slit-like coronary orifice is suggested by the association of this finding with sudden death and ischemic symptoms. Other anatomic features that appear to contribute to the pathophysiology are an acute angle of the takeoff of the coronary, and a long interarterial course. A longer interarterial course, more acute angle of takeoff, and smaller orifice seem to be more common in patients with ALMCA.[13] (B) Histologic specimen of a 22-year-old man who died suddenly during a soccer game. This specimen demonstrates the intramural course of the left main coronary artery as it travels between the aorta (A) and the pulmonary artery (P). When the coronary follows an intramural course, both a slit-like orifice and an acute angle of takeoff are very common. The intramural ALMCA typically arises within the right sinus and has a relatively long intramural course traveling proximal to the sinotubular junction. The intramural ARCA typically arises from higher in the left sinus and travels more distally, at the level of or distal to the intercoronary commissure and sinotubular junction. This feature often obviates the need for significant manipulation of the commissure in an unroofing procedure for ARCA. Note the position of the commissure of the aortic valve in relation to the intramural segment as a potential mechanism of obstruction of this coronary during exercise.

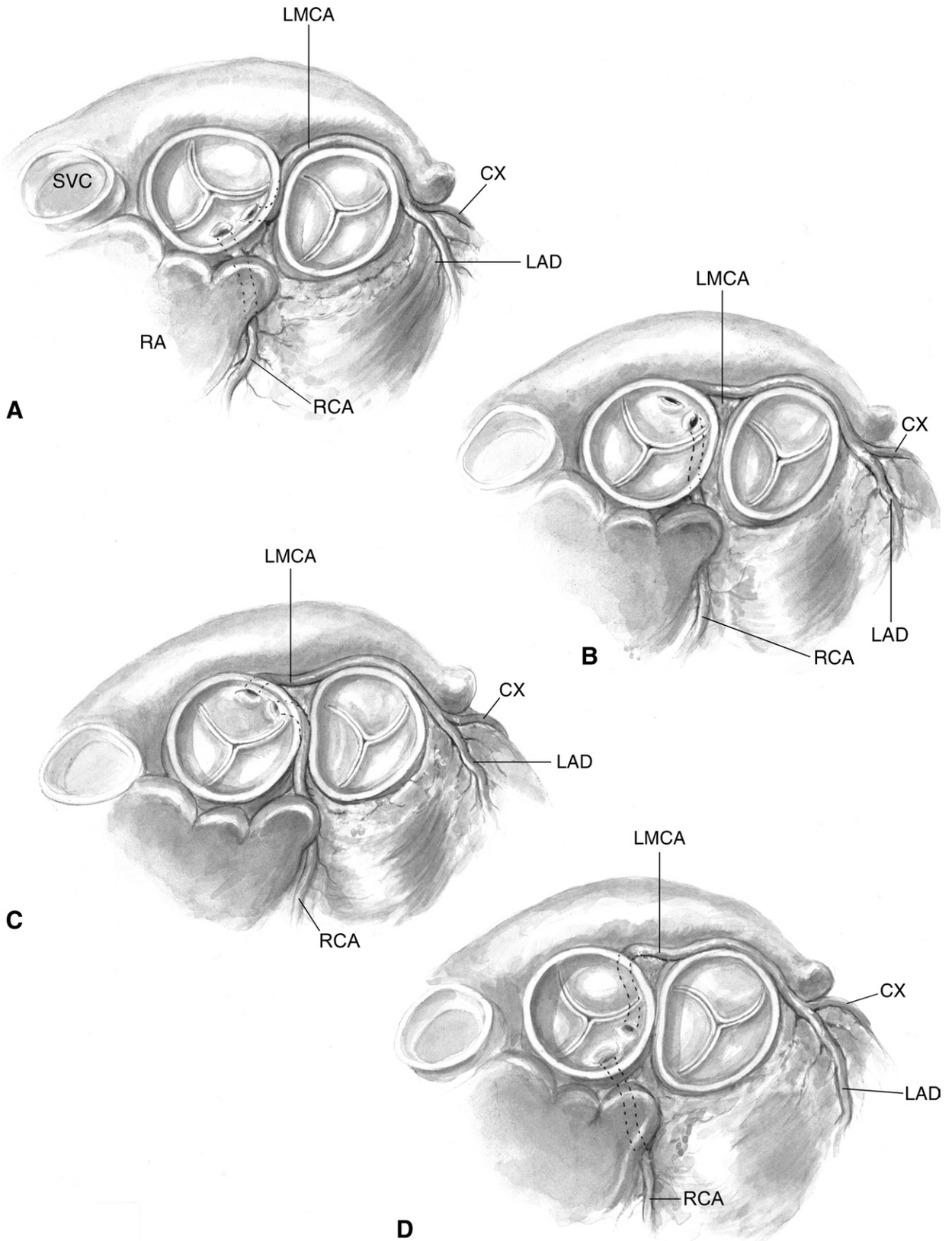


Figure 2 Important surgical anatomy is depicted in this figure. The anomalous coronary may travel between the great vessels in either an extramural (A and C) or an intramural course (B and D). In the case of the intramural coronary, the origin of the coronary may be from either a separate orifice or a common orifice with the opposite coronary. The intramural coronary most often has a slit-like orifice and a very acute of angle takeoff from the lumen of the aorta. In the case of an extramural course, the artery may arise from the aortic sinus, or from the opposite coronary. A slit-like orifice is less common, but an acute angle is often present. It is very important to determine whether the coronary is intramural or extramural before surgical intervention. If this cannot be determined preoperatively, careful dissection must be performed and the unroofing procedure must be avoided in the case of an extramural coronary. CX = circumflex artery; LAD = left anterior descending coronary artery; LMCA = left main coronary artery; RA = right atrium; RCA = right coronary artery; SVC = superior vena cava.

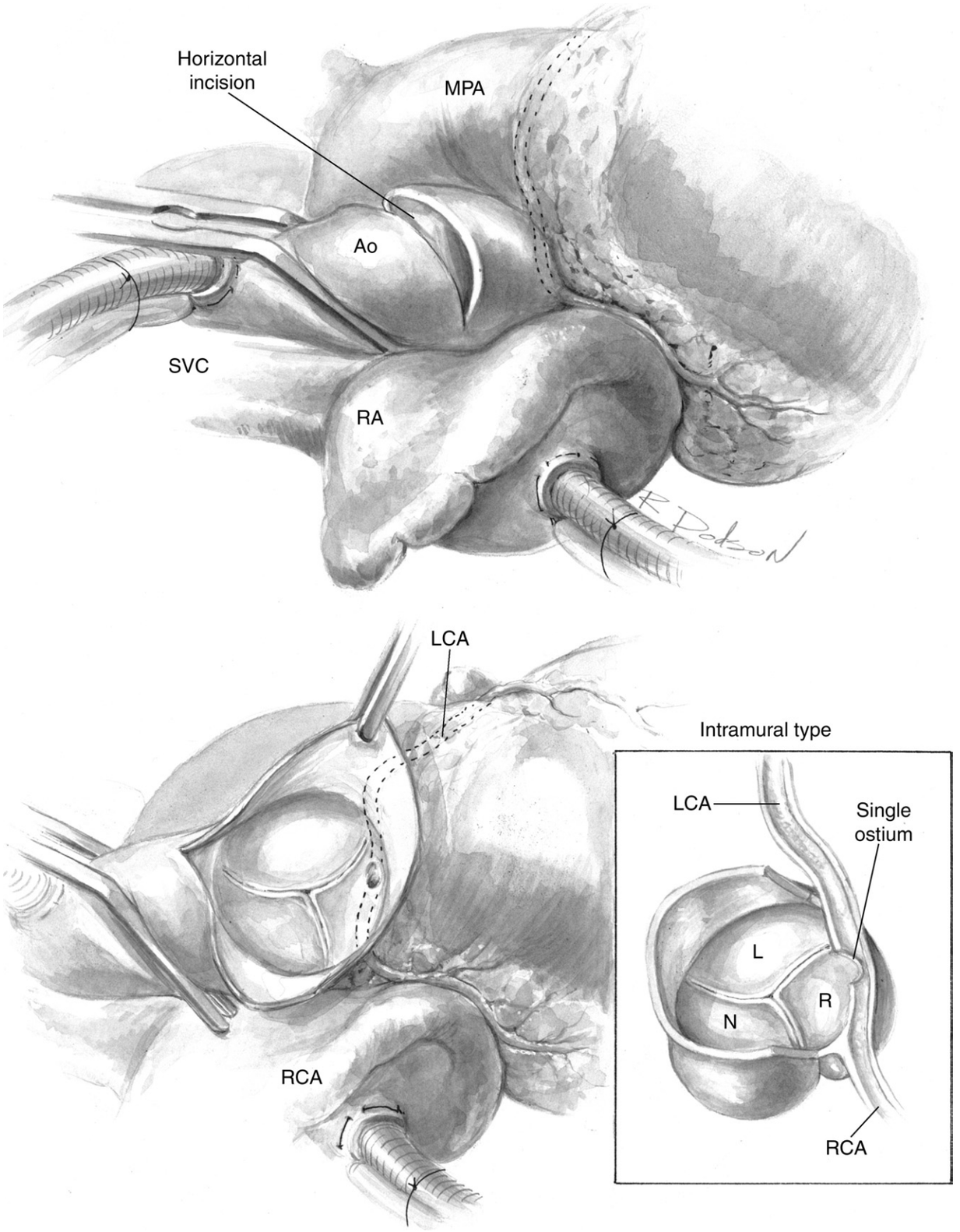


Figure 3 The surgical approach is usually via a median sternotomy but may be accomplished via a limited upper or lower sternotomy. Cardiopulmonary bypass is initiated with distal ascending aortic and right atrial cannulation. An aortic cross-clamp is applied and cardioplegic arrest is initiated. This is typically accomplished with antegrade infusion, but consideration is made for retrograde infusion via the coronary sinus. The space between the great vessels should be mobilized enough to be certain that the artery is intramural. A transverse aortotomy with extension into the noncoronary sinus ("hockey stick") incision is made. Alternatively the aorta may be transected. In either case, great care should be taken to avoid disruption of an intramural course of the coronary with the aortotomy. The anomalous coronary may either have a separate orifice or share a common orifice with the opposite artery. Following aortotomy, the origins of the coronaries are identified, and the course of the intramural segment is gently probed to be sure of its course and the relationship of the coronary to the commissure. Ao = aorta; L = left sinus; LCA = left coronary artery; MPA = main pulmonary artery; N = noncoronary sinus; R = right sinus; RA = right atrium; RCA = right coronary artery; SVC = superior vena cava.

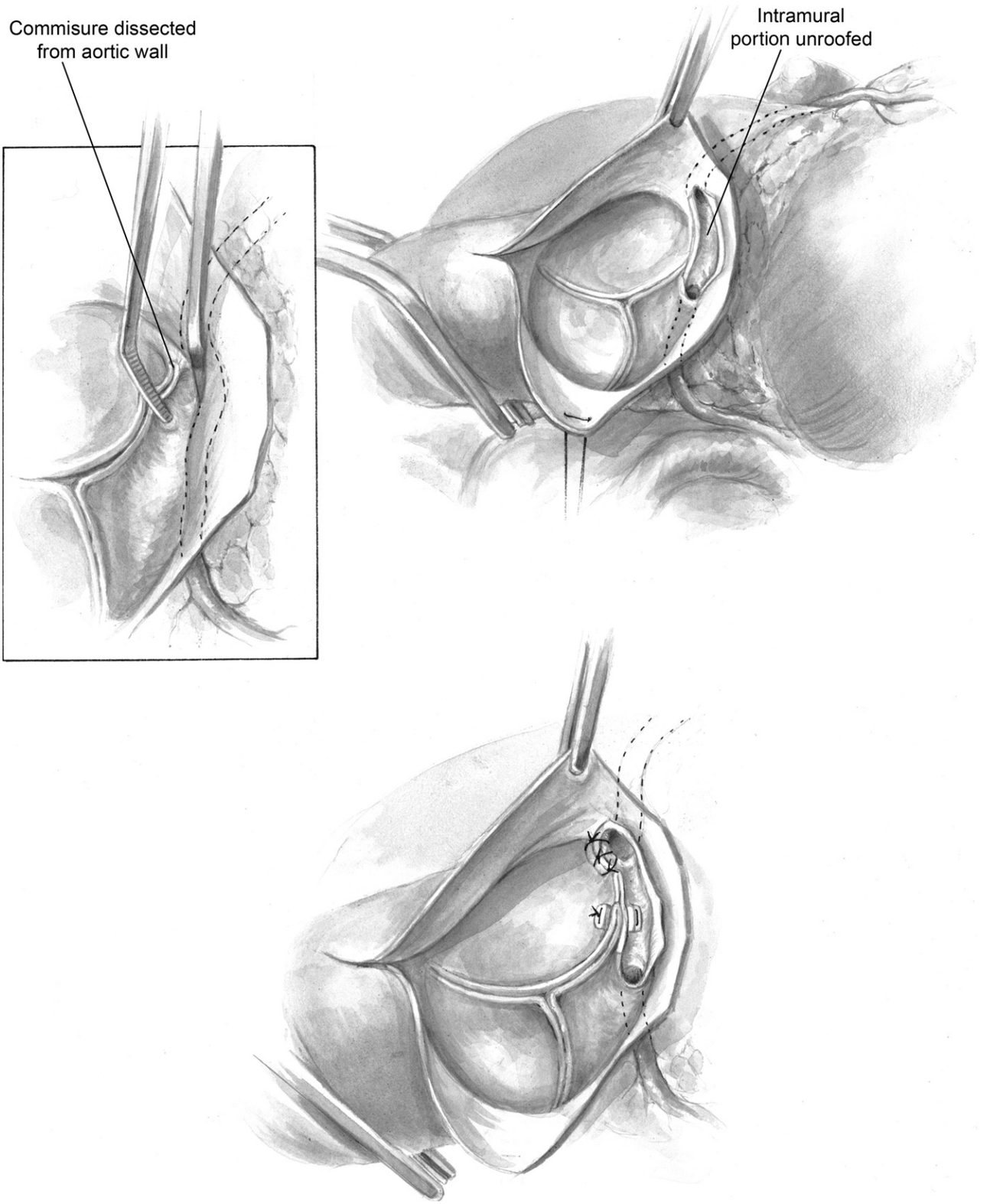


Figure 4 In the modified unroofing procedure, the intramural segment of the anomalous coronary is incised from within the lumen of the aorta up to the point at which the coronary artery leaves the aortic wall in the appropriate sinus. If the origin of the anomalous coronary artery is at a level distal to the commissure, there is very little risk to simply unroofing that segment. In patients in whom the intramural course is at or proximal to the commissure, the commissure may require detachment and reflection into the lumen of the aorta so that unroofing can be accomplished. The commissure should then be secured to the aortic wall at the appropriate level to prevent prolapse of the aortic leaflets and aortic insufficiency. Fine monofilament suture is used to secure any ragged edges of the intima.

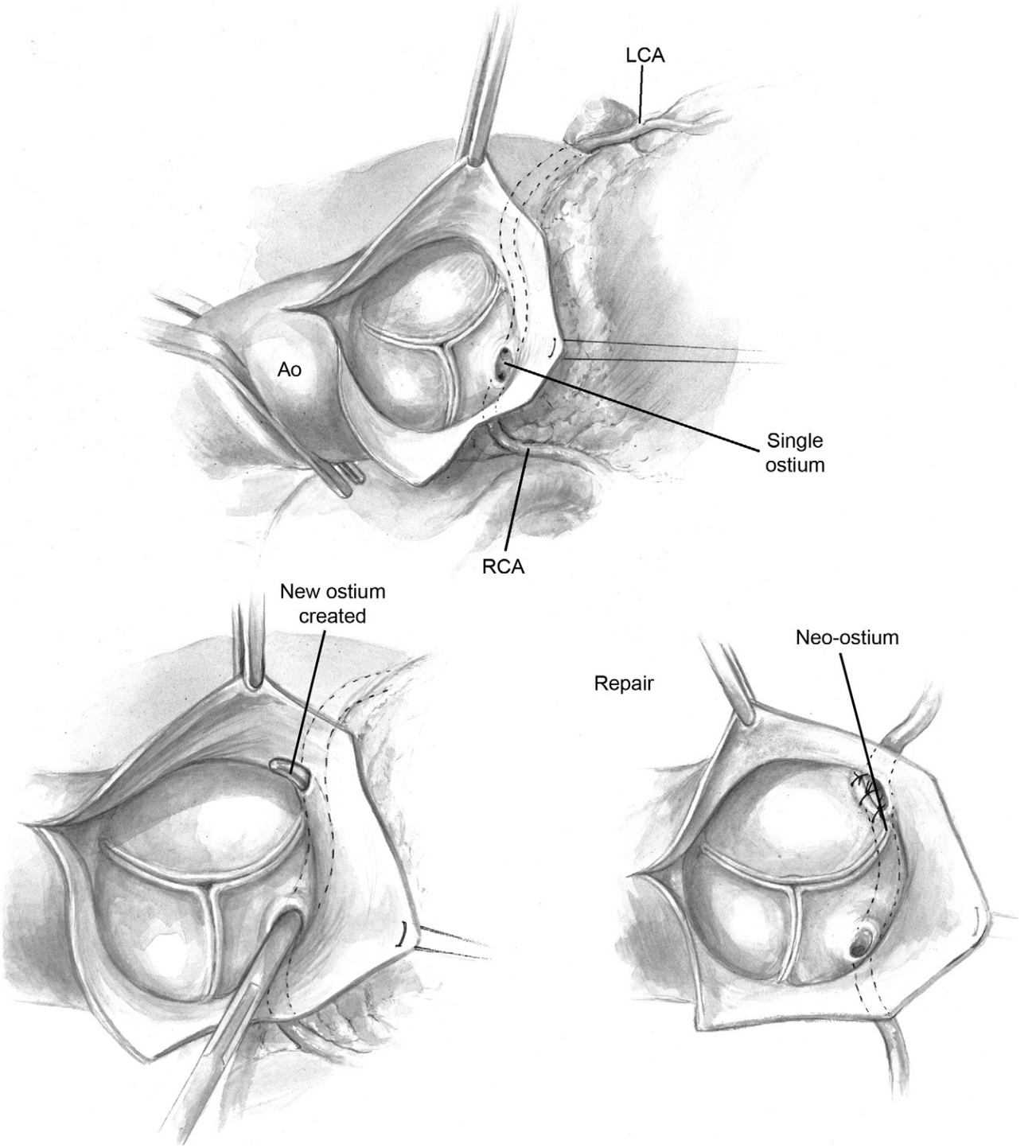


Figure 5 In patients in whom the intramural path of the anomalous coronary artery is at or below the level of the commissure, a neo-ostium can be created by passing a probe or right-angle clamp through the intramural segment to the point at which the coronary artery leaves the aortic wall within the appropriate sinus. A neo-ostium of the coronary is created. Disrupted intima is again secured with fine monofilament suture. It may also be prudent to obliterate the intramural segment of the coronary with a suture to avoid a dual pathway of coronary flow.[22] Ao = aorta; LCA = left coronary artery; RCA = right coronary artery.

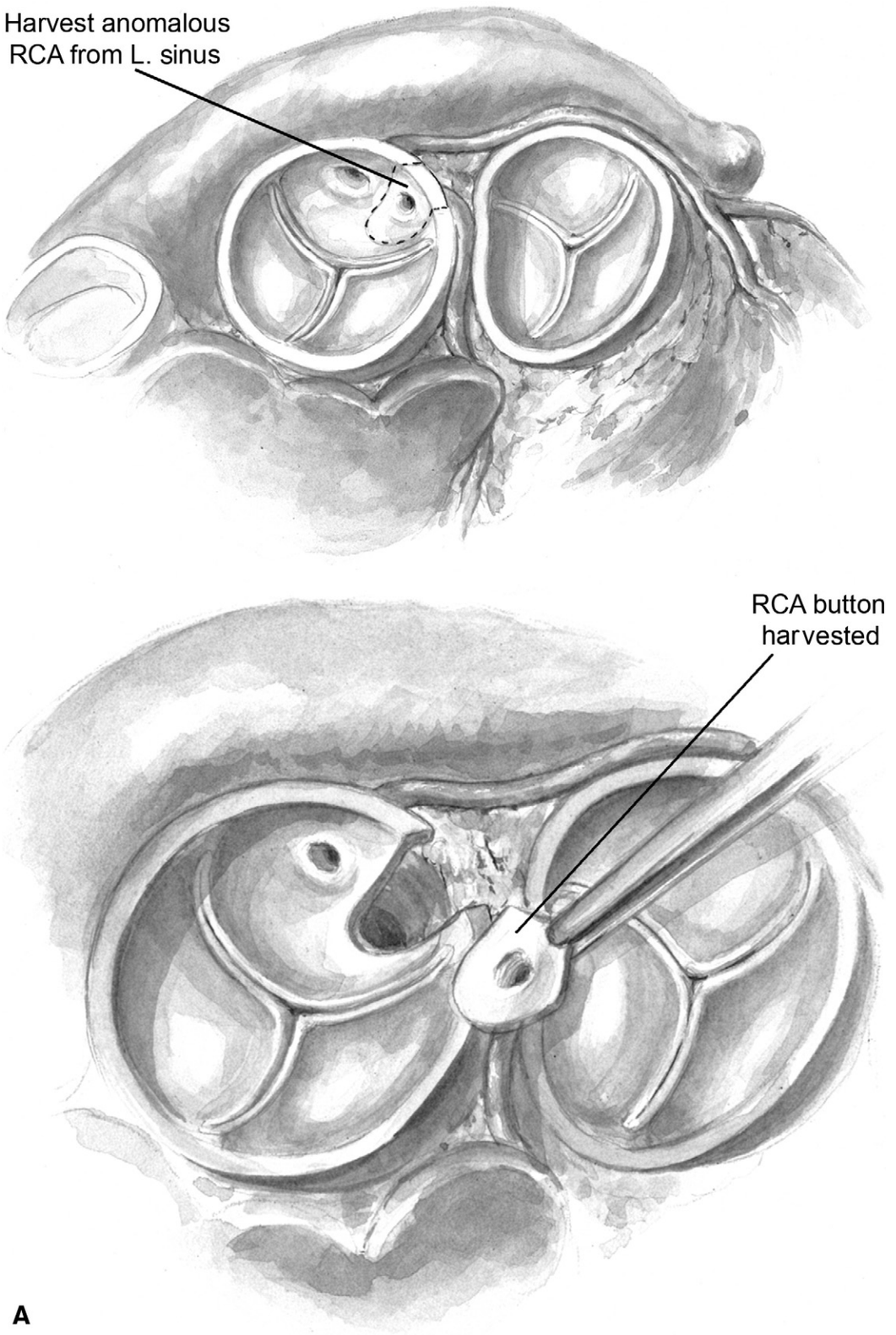


Figure 6 Surgery for patients with an interarterial but extramural course may be individualized. In patients with an extramural course, excision and reimplantation into the appropriate sinus may be the best option. (A) In this technique, the coronary artery must be large enough to transfer directly, or a button of aortic wall must be excised with the coronary artery and implanted in the appropriate sinus. If these options are not possible, CABG with an internal mammary graft may be the most prudent choice. If there is a slit-like orifice of the anomalous coronary, modification of this orifice is necessary to prevent persistent obstruction. (B) The site of the coronary artery harvest is then repaired with prosthetic material (homograft or Dacron), and the coronary is mobilized just enough to be translocated to the appropriate sinus, where it is anastomosed either as a button or in a "V"-shaped incision created in the sinus. It is usually necessary to place this coronary more distal in the sinus or on the aorta to avoid kinking. We routinely use a low-dose nitroglycerine drip for 24 hours to prevent coronary spasm following manipulation. L = left; R = right; RCA = right coronary artery.

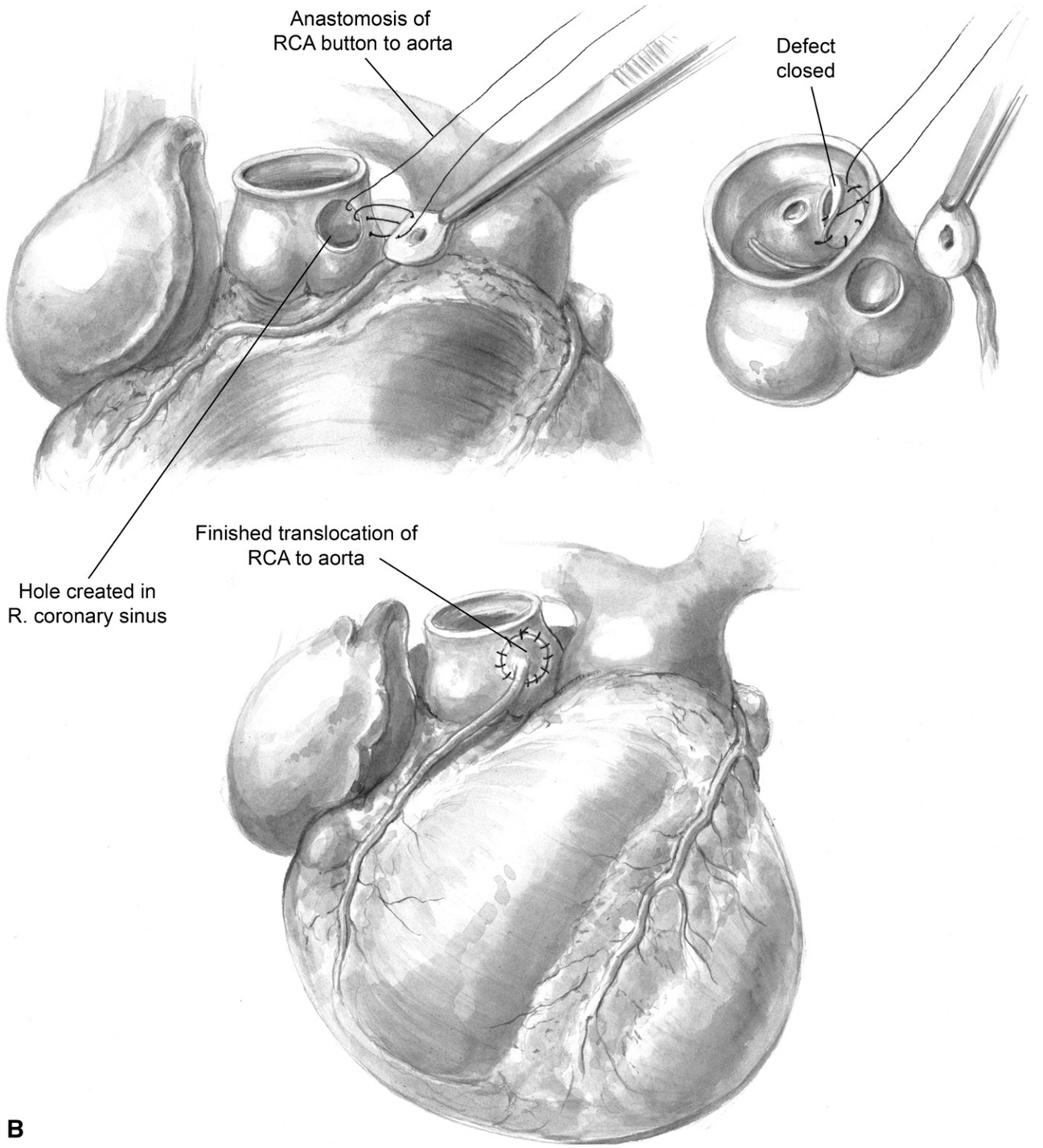


Figure 6 (Continued)

Discussion

Many surgical strategies have been suggested to treat this defect; these include coronary reimplantation, unroofing the intramural segment, and coronary artery bypass grafting (CABG). In most patients with ALMCA and ARCA, correction can be accomplished by either excision and reimplantation[15,16] or a modified unroofing technique.[17] The unroofing procedure has been adopted by many surgeons with good short-term results.

Primary CABG has been advocated because it eliminates the need to open the aorta and manipulate the intercoronary commissure. However, CABG subjects the patient to the complications and potential long-term problems of grafted coronaries. Furthermore, because the flow through the anomalous coronary artery is likely normal at rest, an internal mammary bypass graft may have decreased patency secondary to competitive flow from the native coronary. This has led some authors to recommend ligation of the coronary artery proximal to the insertion of the graft.[18] For this reason, we relegate CABG to older patients with coexistent coronary artery disease. Transcatheter stent placement has also been advocated. This therapy may protect an interarterial coronary from compression but usually does little for the slit-like ori-

fice and may predispose the patient to complications related to the stent.[19] This therapy may be appropriate in an older patient with significant medical problems that would increase the risk of surgery, or with coexistent atherosclerotic coronary artery disease that otherwise would be best treated with angioplasty and stent therapy.

Few data exist regarding the long-term outcome of surgically repaired patients with AAOCA. We have encountered no patient with symptoms referable to ischemia, stenosis, or obstruction following surgery. It is prudent to follow patients treated with either coronary artery reimplantation or unroofing for ostial stenosis or obstruction or, in the case of manipulation of the aortic commissure, for aortic insufficiency. We have reported a series of nine patients treated surgically for anomalous origin of either the left main coronary artery or the right coronary artery. Unobstructed patency of the coronary artery orifice and proximal coronary was demonstrated by echocardiography in eight of nine after repair with modified unroofing procedures; one patient could not be imaged effectively with noninvasive means. All nine patients had a negative stress test or stress echocardiography at a mean of 29 months after repair. We have not recommended routine provocative stress testing following surgery.[20] Since that report, our experience has increased to 30 patients with no mortality or significant complications at a mean follow-up of 29 months.

Conclusions

Anomalous aortic origin of a coronary artery is a relatively rare and potentially lethal anomaly. Patients with an anomalous coronary that follows either an intramural or an extramural course between the great vessels are at risk for cardiac-related events and sudden death. Although the indication for surgery in the asymptomatic patient is controversial, surgery is clearly indicated for the symptomatic patient and can be accomplished with excellent early and mid-term results.

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